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Partial splenectomy in CF patients with hypersplenism

Our recently published article on partial splenectomy in cystic fibrosis (CF) patients with hypersplenism[1] appeared with a commentary by colleagues from the Birmingham Children's Hospital.[2]

The authors of this commentary rightfully point out that liver disease in CF may have a widely varying symptomatology ranging from portal hypertension, bleeding oesophageal varices, ascites, to splenomegaly with hypersplenism. While the quoted clinical experience of 200 patients with CF liver disease might be considered as substantial, it nevertheless appears unjustified to rush from that experience to the statement that severe hypersplenism, requiring a specific surgical approach, is not a feature of the discussed disease spectrum. In the equally substantial clinical experience of our CF centre such severe hypersplenism occurred in only those three patients described in our paper. In these rare cases, however, we found the respiratory and haematological complications caused by the massively enlarged spleens to be impressive and to range from severe impairment of respiratory compliance to life threatening thrombocytopenia. It is for those rare patients that we consider the described surgical intervention to be of potential value. We strongly believe that such patients deserve a therapeutic intervention before their splenomegaly has effected a potentially irreversible deterioration of their chronic lung disease or their hypersplenism has caused a life threatening haemorrhage via thrombocytopenia or infection via leucopenia. The validity of this approach is illustrated by the three reported cases: white blood cell and platelet counts significantly and persistently improved (follow up time is now four to eight years), oesophageal varices disappeared in two and significantly regressed in one patient (reported in our paper and not omitted as postulated in the commentary), and respiratory function improved in all three patients. While the authors of this commentary state that they did not encounter "severe hypersplenism requiring such aggressive management" in their CF patients, one wonders whether early liver transplantation in patients with relatively less severe hypersplenism might not be considered as equally or even more "aggressive".[3]

The reason why white blood cell and platelet counts were not given in our paper was due to the editor's decision to shorten the manuscript.

In contrast to the authors of the commentary we see no reason to believe that liver transplantation and partial splenectomy are surgical interventions that are mutually exclusive. On one side, there are reports of excessive portal hypertension or hypersplenism necessitating splenectomy (or partial splenic embolisation) after liver transplantation,[4–7] on the other side, our surgical colleagues do not see any reason to believe that partial splenectomy actually increases the technical difficulties of a later transplant operation. Furthermore, in admittedly small reported series of partial splenectomies performed in children with a variety of diseases, no major complications have been observed.[8–10]

We agree with the authors of this commentary that liver transplantation, oesophageal band ligation, and transjugular intrahepatic portosystemic stent shunting are important therapeutic options for children with advanced CF liver disease. In contrast to them, however, we welcome partial splenectomy as an additional therapeutic strategy that particularly addresses the problem of splenomegaly and hypersplenism. Ultimately, this difference of perspectives on the same issue might relate to the almost philosophical question whether one welcomes such a new treatment strategy as a potentially promising addition to one's therapeutic quiver, or, alternatively, tends to reject such interesting new possibilities off hand.

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Partial splenectomy—worth the risk

In 1993, we published the results of our first cystic fibrosis (CF) patients who had complications of liver disease and portal hypertension (PHT), and had been operated on by partial splenectomy (PS). We also presented the results of 11 patients at the Jerusalem CF meeting in 1996. Since 1982, we have operated on and followed up 21 patients (aged 8 to 22 years). All patients had a large spleen measuring 15–28 cm in length, oesophageal varices graded 2 to 4 by endoscopy, hypersplenism with a platelet account below 50 000, and a well documented liver disease treated with UDCA.

Surgical procedure consisted of PS with conservation of the upper lobe of the spleen, terminal haemostasis, and suturation of parenchymentous vessels. The whole procedure lasts 3–4 hours. The only postoperative complications consisted of scar rupture in three cases and a painful episode of a few days in two cases. No pulmonary exacerbation occurred after surgery. A speedy normalisation of the haematological profile was observed. Normal function of the remaining upper lobe of the spleen was registered by scintiscan. An important improvement of oesophageal varices was noticed in nine cases out of 11 and a stable condition observed in two cases. The size of the remaining spleen remained stable in nine patients out of the first group who presented in 1996. No deterioration, and even some improvement of hepatic function was observed.

In conclusion, we believe that the risk of PS is worth taking since it appears to be a good option for the treatment of oesophageal varices, which is the main concern, and also might be the cure for hypersplenism. Partial splenectomy is an important alternative to all other procedures in the treatment of PHT. Moreover, it allows a delay of hepatic transplantation and it may even be avoided altogether.

We intend to present our global results in a near future.

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